BRIEF COMMUNICATIONS

RIGHT-SIDED AORTIC ARCH WITH BILATERAL DUCTUS: A RARE CASE OF NONCONFLUENT PULMONARY ARTERIES WITHOUT ASSOCIATED CARDIAC ANOMALIES

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Discontinuity of the pulmonary arteries (PAs) is typically found in conjunction with abnormalities of the pulmonary outflow tract and often in the setting of a right-sided aortic arch or bilateral patent ductus arteriosus (PDA).[1-5] In this lesion, the left and right PA branches originate separately, with one arising from the aortic arch or one of its branches via a PDA and the other from either the pulmonary (or common) trunk or a contralateral PDA. We present a variation of nonconfluent PAs that has not been described previously.

Clinical summary. A term infant was referred at 8 months of age with cyanosis and respiratory distress. Evaluation with echocardiography, magnetic resonance imaging, and cardiac catheterization disclosed normal intracardiac anatomy, a small ostium secundum atrial septal defect, a right-sided aortic arch with an aberrant left subclavian artery arising as the last branch of the arch, and a right-sided PDA from the right PA to the descending aorta. The PAs were discontinuous, with the right branch arising from the pulmonary trunk and the left originating from the proximal segment of the left common carotid artery via an unobstructed left PDA. Pressures in the right ventricle were systemic, with bidirectional shunting through the interatrial defect and the right PDA.

Repair was performed through a median sternotomy, with division of the right PDA, detachment of the left PA and left PDA from the carotid artery, and direct anastomosis of the left PA to the pulmonary trunk. The infant tolerated the procedure well and was discharged 5 days later with no anastomotic stenosis on Doppler evaluation. Follow-up echocardiography 10 years after the operation showed no obstruction to pulmonary outflow or across the anastomotic site, and no intracardiac shunting.

Comment. In their 1966 review of right-sided aortic arch, D'Cruz and associates[6] noted that a right aortic arch with an aberrant left subclavian artery and bilateral PDAs had never been described. In the literature covering the subsequent three decades, we have found but a single case with such a combination, and in that case the left PDA arose from the aberrant left subclavian artery.[3] Bilateral PDAs occur most often in the context of associated heart disease, such as tetralogy of Fallot or anomalies of visceral lateralization (heterotaxy).[3] In fact, one of the circumstances in which bilateral PDAs are found is with discontinuous PAs, with one duct giving rise to the so-called "absent" PA and the other either running between the central PA and systemic arterial systems or, in rare instances with pulmonary atresia, giving sole supply to the contralateral PA branch.[3,4] Similarly rare is the finding of nonconfluent PAs without associated congenital heart defects. Likewise, it is unusual to observe a right-sided aortic arch with an aberrant left subclavian artery and origin of a left PDA from a vessel other than the subclavian artery, or a right-sided aortic arch with a right PDA.[6] All of these conditions were present in our patient.

Insofar as the primordia of the proximal branch PAs are the proximal 6th pharyngeal arches, nonconfluence of the PAs, which presumably results from regression of a proximal 6th arch, is a form of anomalous development of the embryonic arch system (Fig 1). Extrapolating from the primitive pharyngeal arch system, there is a tremendous range of potential anomalies of the aortic arch and its branches, many (but not all) of which have been described.[3,4,7,8] Notably, Freedom and colleagues[3] reported a lesion similar to ours, which differed only in that the left PDA, which also gave rise to a left PA, originated from the aberrant left subclavian artery rather than the left carotid artery. This difference, which might be explained by a different site of junction of the 6th pharyngeal arch with the dorsal aortic system at the time of regression of the proximal 6th arch, suggests an intrinsic developmental heterogeneity of the aortic arch system. The segments of the pharyngeal arch system that regressed in the process leading to the anomalies described in these two cases were likely the same, but the mature anatomy was different, which may indicate differential development before the regression of the proximal 6th arch.

Surgically, this case was straightforward. Reconstruction of PA continuity is a procedure that is not especially uncommon, given the wide range of anomalies in which it is required, including tetralogy of Fallot, pulmonary atresia, truncus arteriosus, and anomalous PA origin from the ascending aorta. As long as the ductus supplying the isolated PA remains patent and the branch PA has been able to develop, reconstruction typically is not problematic. The

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Received for publication Oct 7, 1999; accepted for publication Nov 14, 1999.

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J Thorac Cardiovasc Surg 2000;119:849-51.

0022-5223/2000 $12.00 + 0 **12/54/105459**

doi:10.1067/mtc.2000.105459

The Journal of Thoracic and
Cardiovascular Surgery
April 2000

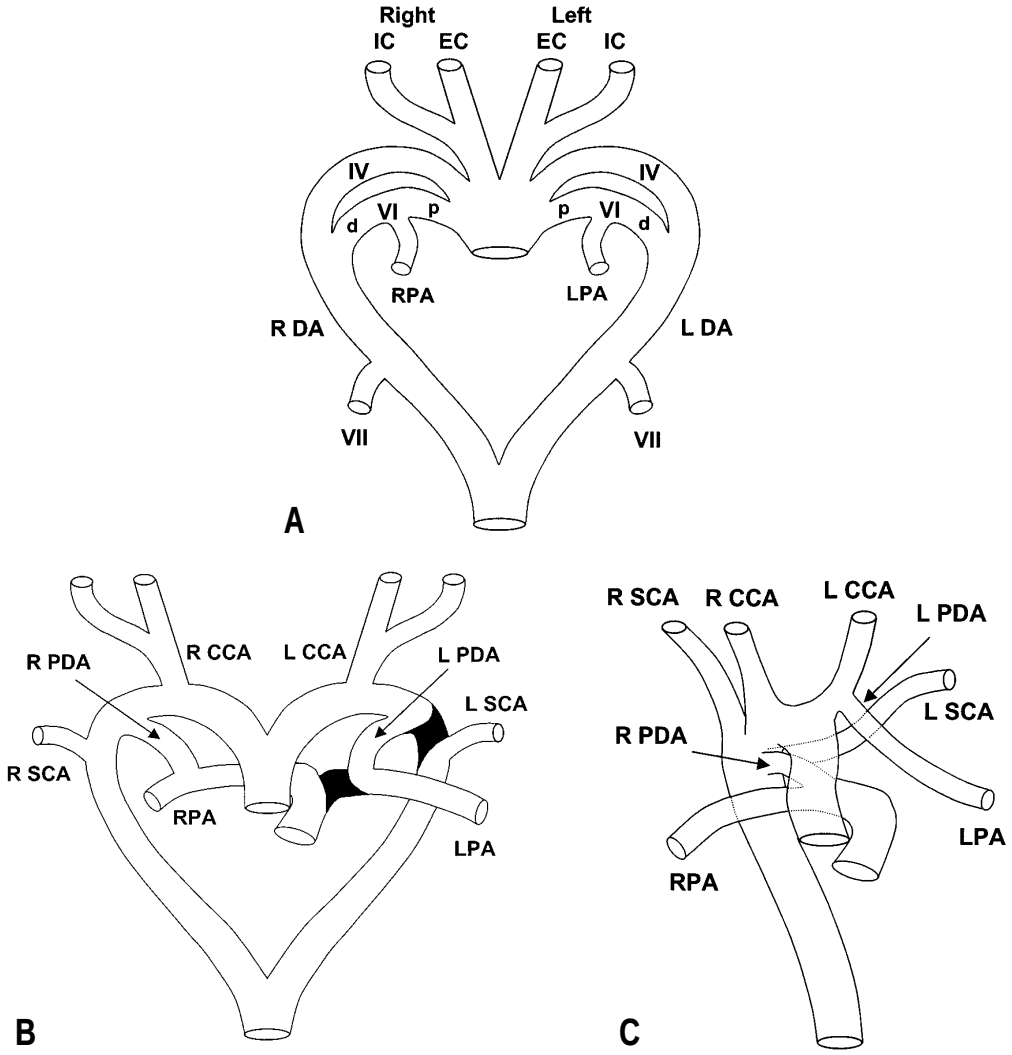

Fig 1. A, Schematic diagram of the primitive pharyngeal arch system, showing the left and right external *(EC)* and internal *(IC)* carotid arteries, 4th *(IV)* and 6th *(VI)* pharyngeal arches, right and left distal pulmonary arterial segments *(PA)*, dorsal aortas *(DA)*, and 7th intersegmental arteries (VII). The proximal *(p)* 6th arches develop into the proximal pulmonary arteries and the distal *(d)* 6th arches become the ductus arteriosus. The 7th intersegmental arteries develop into the subclavian arteries. **B,** Schematic depiction of the regression pattern of the pharyngeal arches that presumably results in the anatomy of our patient. The *black sections* represent arterial segments that regress. *CCA*, Common carotid artery; *L*, left; *PDA*, patent ductus arteriosus; *R*, right; *SCA*, subclavian artery. **C,** Diagram of the mature anatomy of the aortic arch, its branches, and the PAs in our patient.

most likely potential complication is anastomotic PA stenosis. It is important to remember that one PA originates from a PDA, and constriction of ductal tissue may predispose to obstruction, especially in patients in whom the anomaly is repaired early in life. To minimize the potential for stenosis, the surgeon should have a low threshold for patch augmentation across the anastomosis. Postoperatively, close follow-up is indicated to detect stenosis that does develop before it becomes symptomatic, at which point irreparable damage may have already occurred to the distal pulmonary vascular bed. Stenosis will most likely become manifest during the first year or so after repair, as the process of remodeling takes place. Stenosis may become more prominent as the patient enters a growth phase, and follow-up should be continued at least through adolescence in patients who have undergone reconstruction of nonconfluent PAs in infancy or childhood.

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